Pregnancy Among Women With Congenitally Corrected Transposition of Great Arteries

Heidi M. Connolly, MD, FACC, Martha Grogan, MD, FACC, Carole A. Warnes, MD, MRCP, FACC
Rochester, Minnesota

OBJECTIVES
The outcome of pregnancy in congenitally corrected transposition of the great vessels was studied in 22 women.

BACKGROUND
Women with congenitally corrected transposition of the great vessels often reach childbearing age. Although reports on the outcome of pregnancy in these women are available, the number of patients is small.

METHODS
The medical and surgical databases at the Mayo Clinic were reviewed, and 36 women >16 years old with congenitally corrected transposition of the great vessels were identified. All of them were contacted, and 22 who had pregnancies were identified and the outcome of pregnancy was evaluated.

RESULTS
Twenty-two women had 60 pregnancies resulting in 50 live births (83%). Forty-four deliveries (88%) were vaginal and 6 (12%) were by cesarean section. One delivery was premature at 29 weeks. There was one successful twin pregnancy. There were 11 unsuccessful pregnancies. One patient developed congestive heart failure late in pregnancy because of systemic atrioventricular valve regurgitation and required valve replacement in the early postpartum period. One patient had a total of 12 pregnancies, including 1 twin pregnancy and 2 unsuccessful pregnancies. She had multiple pregnancy-related complications, including toxemia, congestive heart failure, endocarditis and myocardial infarction (single coronary artery). No other serious pregnancy-related maternal complications and no pregnancy-related deaths occurred. The mean birth weight of the infants (n = 32) was 3.2 ± 0.4 kg. None of the 50 live offspring have been diagnosed with congenital heart disease.

CONCLUSIONS
Successful pregnancy can be achieved in most women with congenitally corrected transposition of the great arteries. The rate of fetal loss and maternal cardiovascular morbidity is increased. Because of the small number of births, the risk of congenital heart disease in offspring of women with congenitally corrected transposition of the great arteries is uncertain. (J Am Coll Cardiol 1999;33:1692–5) © 1999 by the American College of Cardiology

Congenitally corrected transposition of the great arteries (l-TGA) is an uncommon form of congenital heart disease that accounts for <1% of all congenital cardiac defects (1) and is characterized by atrioventricular and ventricular–great arterial discordance. Although l-TGA may occur as an isolated congenital anomaly (2), it usually is associated with other congenital abnormalities (3,4), including ventricular septal defect, pulmonary stenosis (valvular or subvalvular), anomalies of the atrioventricular valves (Ebstein-like malformation of the systemic atrioventricular valve), complete heart block, dextrocardia and (less commonly) atrial septal defect. Often, patients with isolated l-TGA are initially identified during adult life. Survival into adulthood is common, either after operation (5,6) or with isolated l-TGA. From 30% to 50% of patients with l-TGA have systemic ventricular dysfunction and systemic atrioventricular valve regurgitation, because the morphologic right ventricle and tricuspid valve are in the systemic circulation (7,8).

The outcome of pregnancy in l-TGA has been described (2,9,10), but no pregnancy series has been reported in such patients. To better define the maternal and fetal outcome of pregnancy in women with congenitally corrected transposition of the great arteries is uncertain. (J Am Coll Cardiol 1999;33:1692–5) © 1999 by the American College of Cardiology

METHODS
The medical and surgical databases of the Mayo Clinic were reviewed, and 36 women of childbearing age (>16 years) with l-TGA who were evaluated at the Mayo Clinic in the last 17 years (1980 to 1997) were identified. The patients were evaluated either in follow-up by one of the authors or contacted through the mail or by telephone. Their medical records were reviewed. Twenty-two women who had pregnancies were identified. A detailed obstetric history that
included details of menstrual history, pregnancy, delivery and fetal outcome was obtained from each woman. Correspondence from local obstetricians and cardiologists was reviewed; however, these physicians were not contacted directly.

The clinical, surgical and diagnostic (electrocardiography, chest radiography and echocardiography) data were reviewed for each patient. All patients had the diagnosis of l-TGA confirmed by one or more of the following: comprehensive echocardiographic examination, cardiac catheterization and surgical intervention. Congenital cardiac surgery was performed in nine women before pregnancy and in eight women after pregnancy.

Fourteen women with l-TGA have not had a pregnancy and in none of them has pregnancy been contraindicated. Eight women who were advised against pregnancy subsequently became pregnant. They had 11 pregnancies, and 10 were successful. None of the women in the present series reported infertility.

Statistical methods. Pregnancy and birth weight variables were compared between women who previously had surgical intervention for l-TGA and those who had not. Mean values ± standard deviations were calculated for continuous variables and evaluated for statistical significance by a two-sample t test. Absolute and relative frequencies were measured for discrete variables and evaluated with the Fisher exact test. A p value < 0.05 was considered statistically significant.

RESULTS

Twenty-two women with l-TGA had 60 pregnancies (Fig. 1). The mean maternal age at the time of delivery was 28.6 ± 5.6 years (range, 19 years to 45 years). Four women had isolated l-TGA and 18 had associated anomalies. Associated cardiac anomalies included ventricular septal defect in 10 women, pulmonary valve or subvalvular stenosis in 7, atrial septal defect or patent foramen ovale in 6, dextrocardia in 5, and straddling systemic atrioventricular valve, Ebstein-like systemic atrioventricular valve and a single coronary artery in one each. Congenital heart disease has not been identified in any of the live offspring. The unsuccessful pregnancies were not evaluated for congenital anomalies.

Of the 22 women, 9 had 1 or more cardiovascular procedures (4 had reoperation) before pregnancy. All of these women had significant coexistent cardiovascular disease. The procedures performed included ventricular septal defect closure in 7 women, conduit placement in 5 (1 conduit re-replacement), atrial septal defect closure in 3, systemic atrioventricular valve replacement in 2, Blalock-Taussig shunt placement in 2, and pulmonary artery band and permanent pacemaker implantation in 1 each.

After pregnancy, 10 women had 1 or more cardiovascular procedures. Two operations were performed within 6 months after delivery; these included 1 atrioventricular valve replacement and 1 atrial septal defect closure. Other interventions were performed a mean of 16 years after pregnancy (range, 2 months to 41 years) and included valve replacement, atrial septal defect closure, permanent pacemaker implantation, ventricular septal defect closure, percutaneous transluminal coronary angioplasty and cardiac transplantation.

Pregnancy outcome. Overall, 50 of the 60 pregnancies (83%) were successful, including one twin pregnancy. There were 8 miscarriages (first trimester), two spontaneous abortions (second trimester), and 1 stillbirth in 6 women. Of the 50 deliveries, 44 (88%) were vaginal and 6 (12%) were by cesarean section (Fig. 1). Three of the cesarean sections were performed for obstetric reasons and the other three for perceived maternal cardiovascular risk. There were no pregnancy-related maternal deaths; however, one woman developed congestive heart failure late during pregnancy because of systemic atrioventricular valve regurgitation (ejection fraction, 45%). This patient required atrioventricular valve replacement two months’ postpartum. One patient developed hypertension and gestational diabetes during pregnancy. Another woman, who had 12 pregnancies, had multiple pregnancy-related complications, including congestive heart failure, toxemia, endocarditis and a myocardial infarction (single coronary artery).

The mean birth weight of 32 infants born to women with l-TGA was 3.2 ± 0.4 kg. The mean birth weight of nine infants born to women who had undergone a previous cardiac operation was significantly lower than that of 23 infants born to women who had no previous cardiac operation (2.8 ± 0.5 kg vs. 3.3 ± 0.4 kg) (p = 0.006). Birth weights were not available for 18 infants.

Pregnancy in women with a previous cardiovascular procedure. Nine women had pregnancies after one or more cardiovascular procedures for l-TGA and associated anomalies. There were 29 pregnancies, 27 of which were successful. Seven women had previous ventricular septal defect closure, 5 had simultaneous conduit placement from the right ventricle to the pulmonary artery, and 1 had conduit replacement for conduit obstruction. Three women had atrial septal defects closed, 2 had systemic atrioventricular

Figure 1. Results of 60 pregnancies in women with congenitally corrected transposition of the great vessels. There were no therapeutic abortions.
valves replaced (1 Ionescu-Shiley and 1 Carpentier-Edwards), 2 had a prior Blalock-Taussig shunt, 1 had a pulmonary artery band and 1 had a permanent pacemaker implanted before pregnancy. There was no evidence of premature degeneration of the prosthetic atrioventricular valves in either patient (follow-up is currently at 3 and 13 years after implantation).

**Premature births.** There was 1 premature birth (before 38 weeks) among 50 successful pregnancies. The premature birth occurred at 29 weeks in a woman who had previously had cardiac surgery (l-TGA associated with ventricular septal defect, valvular and subvalvular pulmonary stenosis, type C straddling of the left atrioventricular valve) and was due to placenta previa.

**Unsuccessful pregnancies.** Of the 60 pregnancies in women with l-TGA, 11 were unsuccessful, including 8 miscarriages, 2 spontaneous abortions and 1 stillbirth. The 8 miscarriages occurred in 4 women without previous cardiac surgery: 2 women subsequently had surgical intervention and 2 still have not had an operation. One miscarriage occurred in an aycanotic patient with two previous operations: initial Blalock-Taussig shunt and subsequent ventricular septal defect closure and right ventricle-to-pulmonary artery conduit placement. This patient had atrial septal defect closure after pregnancy. One spontaneous abortion occurred in a patient with two previous cardiac operations: initial ventricular septal defect closure and conduit placement from the right ventricle to the pulmonary artery. The premature births occurred in patients in whom the diagnosis of l-TGA had not yet been made at the time pregnancy occurred.

**DISCUSSION**

Significant hemodynamic and physiologic changes occur during pregnancy (11), some of which may adversely affect the hemodynamic status of a woman with uncorrected significant congenital heart disease such as l-TGA. These changes include an average increase in cardiac output of 40% and an increase in stroke volume, heart rate and concentration of catecholamines in the circulation. A decrease in peripheral vascular resistance is also noted.

Few reports of pregnancy are available about women with l-TGA (2,9). In the present series, pregnancy was well-tolerated in all but two women. One woman required atrioventricular valve replacement in the early postpartum period. The second had congestive heart failure during three pregnancies and toxemia, endocarditis and a myocardial infarction during three other pregnancies. She is alive at age 80 years and recently had percutaneous transluminal coronary angioplasty but no other cardiac surgical intervention.

The rate of cesarean section among the study group was 12% (6 of 50 deliveries), which is lower than the standard rate of up to 23.5% (National Center for Health Statistics, 1990). The miscarriage rate was 16%, which is somewhat higher than the expected rate of 10% (12). There was one stillbirth and two spontaneous abortions. No therapeutic abortions were performed.

Atrioventricular valve regurgitation and ventricular dysfunction are recognized important complications in patients with l-TGA, because the morphologic right ventricle and tricuspid valve are in the systemic circulation (2–8). Among the women in the present series, one developed symptomatic atrioventricular valve regurgitation that required surgical intervention in the early postpartum period. The diagnosis of l-TGA was first made when the patient went into congestive heart failure postpartum. Another woman had multiple pregnancy-related complications, including congestive heart failure, endocarditis, toxemia and myocardial infarction. One woman developed systemic hypertension and gestational diabetes during pregnancy, and one had a premature delivery. The significant difference in birth weight between the offspring of women who had been operated on and those who had not may reflect the lack of birth weight data for 18 offspring or differences in the severity of the disease. None of the women in this series were cyanotic during pregnancy.

**Study limitations.** Although the present report is the largest series describing the outcome of pregnancy in women with l-TGA, there are several limitations. The study is retrospective and subject to referral bias. Obstetric details were obtained directly from each patient and correspondence was reviewed; the obstetricians were not contacted directly; and birth weights were not available for approximately one-third of the offspring. The degree of ventricular dysfunction and atrioventricular valve regurgitation was not evaluated at this institution during pregnancy. Finally, because of the small number of offspring of patients with l-TGA and the lack of formal examination, the incidence of congenital heart disease in the offspring of patients with l-TGA remains undetermined.

**Conclusions.** Most women with l-TGA reach childbearing age. Appropriate evaluation, assessment and recommendations need to be made when these patients wish to pursue pregnancy. A comprehensive cardiovascular evaluation, including an evaluation of cardiac rhythm, systemic ventricular function, atrioventricular valve function, associated lesions and postoperative sequelae, should be performed. Women with New York Heart Association functional class III or class IV symptoms, significant systemic ventricular dysfunction (ejection fraction <40%) or significant systemic atrioventricular valve regurgitation should be counseled against pregnancy.

It is recommended that a pregnant woman with l-TGA be monitored closely by a multidisciplinary health care team that includes obstetricians, cardiologists and obstetric anesthesiologists. Care must be taken to avoid rapid volume shifts during pregnancy and parturition. High-risk patients should be delivered in a monitored setting with Swan-Ganz catheterism.
catheterization if appropriate. The second stage of labor should be facilitated to decrease the excessive hemodynamic stress on the patient. Endocarditis prophylaxis should be administered to high-risk patients (13).

The results of this study suggest that l-TGA does not inhibit fertility in women before or after surgical repair. Successful pregnancy can be achieved in most women, although there is increased risk of maternal cardiovascular morbidity and fetal loss.

Reprint requests and correspondence: Dr. Heidi M. Connolly, Mayo Clinic, 200 First Street SW, Rochester, Minnesota 55905.

REFERENCES